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Letter to the Editor: "Response to Hord and Janco re Chemotherapy for Unresectable Pancreatoblastoma"

We read with interest Hord and Janco's letter on the treatment of unresectable pancreatoblastoma [1]. Their patient showed no response to combination chemotherapy and died 6 months after diagnosis. As this is a rare childhood malignancy, we would like to report our patient, who nevertheless showed some response to combination treatment with vincristine, dactinomycin, and ifosfamide.

A 3-year-old Chinese boy presented with progressive abdominal distention and diarrhea for 10 days. An enlarged liver was palpable 5 cm below the costal margin. On computed tomography (CT) scan, a large hypodense mass (5 cm × 7 cm) with heterogeneous attenuation and calcified speckles was seen anterior to the left kidney and posterior to the stomach, compatible with a pancreatic tumor. Multiple hepatic metastases were present. Skeletal survey was negative. The serum α-fetoprotein (AFP) was 59.7 ng/ml. The urinary catecholamines were not increased, and the bone marrow was clear of malignant infiltrate. Biopsies taken from the primary tumor and the liver confirmed the diagnosis of metastatic pancreatoblastoma.

The child was treated with vincristine 1.5 mg/m^2 on days 1 and 8, ifosfamide 3 mg/m^2 on days 1 and 2, and dactinomycin 0.9 mg/m^2 days 1 and 2. The course was repeated every 3 weeks when neutropenia recovered. A transient response was evident between the second and sixth months after diagnosis when the serum AFP returned to normal levels (Fig. 1). The follow-up CT scan performed at 4 months revealed significant reduction in the primary tumor with maximum dimensions measuring 3 cm × 5 cm. However, the tumor became progressively refractory to chemotherapy. The patient finally died 16 months after diagnosis.

The prognosis of children with disseminated pancreatoblastoma remains dismal. The use of chemotherapy in such cases is largely palliative, and a reasonable symptom-free survival can be attained in a few patients [2,3]. Definitive treatment relies on surgery, although preoperative chemotherapy may be useful [4]. Given the paucity of reported cases, we are still not sure of the optimal chemotherapeutic regimen. The present case may contribute to the search for an effective combination of chemotherapy in the treatment of this rare malignancy of childhood.

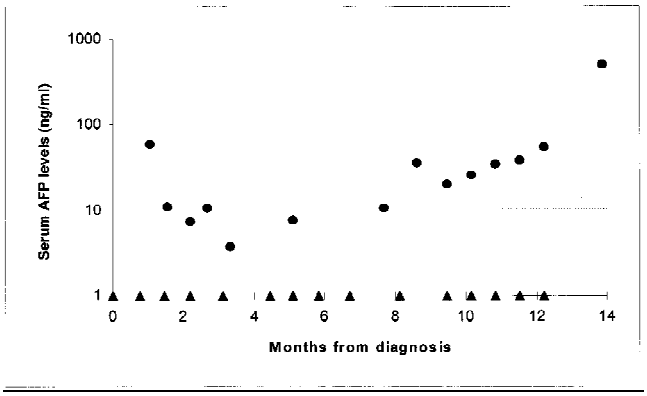

Fig. 1. Changes in the serum α-fetoprotein (AFP) levels (●) in relation to the pulses of chemotherapy (▲) during the course of illness.

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